

Definitions versus categorisation: Assessing the development of lexico-semantic knowledge in Williams syndrome

Harry R. M. Purser¹, Michael S. C. Thomas¹, Sarah Snoxall², Denis Mareschal² & Annette Karmiloff-Smith¹

¹Developmental Neurocognition Lab, Birkbeck, University of London, UK

²Centre for Brain and Cognitive Development, School of Psychology, Birkbeck College, London, UK

Running head: Lexical knowledge in WS

Word count: 6,000

Address for correspondence:

Dr. Harry Purser
Developmental Neurocognition Lab
School of Psychology
Birkbeck College
Malet Street
London WC1E 7HX, UK
Email: h.purser@bbk.ac.uk
Web: <http://www.psyc.bbk.ac.uk/research/DNL/>
Tel.: +44 (0)20 7631 6386
Fax: +44 (0)20 7631 6312

Abstract

Williams syndrome (WS) is associated with relatively strong language abilities despite mild to moderate intellectual disability, particularly when language is indexed by vocabulary. The aim of the study was twofold: (1) to investigate whether reported lexical anomalies in WS can be explained with reference to anomalous semantic development; (2) to assess whether receptive vocabulary skills in WS, a relative strength, are underpinned by commensurate semantic knowledge. The development of lexical-semantic knowledge was investigated in 45 typically developing (TD) individuals (CA range: 5-10, MA range: 5-13) and 15 individuals with WS (CA range: 12-50, MA range: 4-17), by means of (a) a categorisation task and (b) a definitions task, which was expected to make additional metacognitive demands. At younger ages, the performance level of TD individuals and individuals with WS did not differ on the definitions task. However, the WS group's ability to define words fell away from the level predicted by the TD group at older ages, as more sophisticated definitions were expected. The results of the categorisation task indicated that individuals with WS had less lexical-semantic knowledge than expected given their level of receptive vocabulary, although from this lower level, the knowledge then developed at a similar rate to that found in typical development. We conclude first that conventional vocabulary measures may overestimate lexical-semantic knowledge in WS, and second concerns about the metacognitive demands of the definitions task when used with atypical populations may be well founded (cf. Benelli et al., 1988).

Abbreviations: Williams syndrome (WS), typically developing (TD), chronological age (CA), mental age (MA), verbal mental age (VMA), British Picture Vocabulary Scale (BPVS)

Introduction

The uneven cognitive profiles presented by various developmental disorders are often used to investigate different aspects of cognition and to inform theories of cognitive development. Williams syndrome (WS) is a rare neurodevelopmental disorder, with a prevalence estimated to be about 1 in 20,000 (Morris, Dempsey, Leonard, Dilts & Blackburn, 1988), resulting from a microdeletion of more than 25 genes from one copy of chromosome 7 (Ewart et al., 1993, see Donnai & Karmiloff-Smith, 2000). The disorder is characterised by an uneven cognitive profile: despite particular difficulties with visuospatial and numerical cognition, language abilities appear less impaired in individuals with WS (Ansari, Donlan, Thomas, Ewing, Peen & Karmiloff-Smith, 2003; Donnai & Karmiloff-Smith, 2000; Farran & Jarrold, 2003; Howlin, Davies & Udwin, 1998). Moreover, some researchers have claimed that language is ‘selectively preserved’ in WS (e.g. Bellugi et al., 1990; Bellugi, Lichtenberger, Jones, Lai & St. George, 2000), and have used the apparent disparity between language and other areas of cognitive ability as evidence that language develops independently (as a module) from those other cognitive abilities (e.g. Anderson, 1998; Bellugi, Marks, Bihle & Sabo, 1988; Rossen, Bihle, Klima, Bellugi, & Jones, 1996). Although such claims would clearly be of great theoretical importance if correct, their validity has been questioned by other researchers.

In a theoretical objection, Thomas and colleagues (Thomas, Dockrell, Messer, Parmigiani, Ansari & Karmiloff-Smith, 2006) pointed out that the logic of such claims is based on the assumption that deficits in the current state of a cognitive system directly reflect the conditions of the initial state, ignoring the role of development in determining the current state (see Thomas, Purser & Richardson, in press, for discussion). An empirical objection to claims of a language advantage in WS was made by Brock (2007), who reviewed existing studies and concluded that there is, in fact, little evidence that morphology, phonology,

pragmatics, or syntax are better than predicted by nonverbal measures. An alternative (and perhaps more conservative) view, then, is that the cognitive profile associated with WS reflects the outcome of atypical development, with multiple interacting constraints, that has stronger effects on some cognitive abilities and weaker effects on others (Karmiloff-Smith, 1998). Nevertheless, although delayed, language development in WS shows many similarities to typical language development, albeit with some noted exceptions in early precursors (Thomas, 2008; Thomas & Karmiloff-Smith, 2005).

Although younger children with WS have receptive vocabularies in line with their verbal mental age (Thal, Bates, & Bellugi, 1989; Volterra, Caselli, Capirci, Tonucci, & Vicari, 2003), this area of language appears to be a relative strength for older children and adults with the disorder (e.g. Brock, Jarrold, Farran, Laws, & Riby, 2007; Rossen et al., 1996). Indeed, one notable feature of vocabulary in WS is the reported prevalence of rare or low-frequency words in discourse (Bellugi, Wang, & Jernigan, 1994; Udwin & Dennis, 1995). These unusual choices of words can contain unnecessary, or even inappropriate, contextual details (Rossen et al., 1996), leading the speech of individuals with WS to be described as displaying ‘cocktail party syndrome’, referring to the tendency to talk at length with only superficial understanding (Bellugi, Birchle, Neville, Jernigan, & Doherty, 1992).

This phenomenon has led to the suggestion that the lexicon is atypical in WS. For example, Rossen et al. (1996) investigated how individuals with WS interpret homonyms, such as ‘bank’. It was found that the 10- to 18-year-old participants with WS interpreted homonyms with the secondary meaning (‘river’) as often as the first meaning (‘place that stores money’) and more often than both individuals with Down syndrome (DS), matched on both mental age (MA) and chronological age (CA), and typically-developing 10-year-olds. Rossen and colleagues suggested that these group differences resulted from atypical inhibitory processes

failing to integrate contextual information in the lexicon. Temple, Almazan, and Sherwood (2002) have also argued for an atypical lexicon in WS, drawing evidence from a word-picture matching task adapted from the British Picture Vocabulary Scale (BPVS: Dunn, Dunn, Whetton, & Burley, 1997), but involving fine-grained semantically-related distractors. Compared to a typically developing control group matched on MA, their participants with WS performed poorly, leading Temple and colleagues to conclude that individuals with WS have difficulties storing or activating semantic information in the lexicon.

However, many studies have cast doubt on suggestions of atypical lexical structure in WS. In verbal short-term memory tasks, individuals with WS have demonstrated similar semantic effects to control groups matched on receptive vocabulary and digit span, with superior recall for concrete over abstract words (Laing, Grant, Thomas, Parmigiani, Ewing, & Karmiloff-Smith, 2005). Moreover, Tyler and colleagues (Tyler, Karmiloff-Smith, Voice, Stevens, Grant, Udwin, Davies, & Howlin, 1997) found normal sensitivity to semantic priming in a WS group: responses were quicker to words preceded by either a functionally-related word or a category-related word than when preceded by an unrelated word. Furthermore, Bellugi and colleagues found no differences between a WS group and an age- and IQ-matched group with DS on a word definition task (Bellugi et al., 1990).

Thomas and colleagues investigated possible atypical dynamics in lexical access in WS (Thomas et al., 2006), using a speeded picture-naming task in which frequency and semantic category were manipulated as implicit variables. Although the participants with WS were slower and less accurate than a control group matched on receptive vocabulary, the frequency and semantic category effects shown by the participants with WS were found to be in line with controls, indicating normal encoding of frequency in the WS lexicon and a similar categorical structure.

Further evidence has been gained from semantic fluency tasks. Despite Bellugi and colleagues' widely-cited finding that a WS group produced more atypical responses in a semantic fluency task than a DS group when the semantic category was animals (Bellugi et al., 1990), the authors did not present any statistical analyses of the typicality of response. Subsequent investigations of semantic fluency in WS have largely failed to find group differences in typicality or frequency of verbal output (e.g. Jarrold, Hartley, Phillips, & Baddeley, 2000; Johnson & Carey, 1998; but see Temple et al., 2002). However, the possibility remains that metacognitive demands in such tasks, such as retrieval strategies and evaluation of task completion, might cloud our view of the underlying structure of the WS lexicon; individuals with WS might reveal an atypical lexicon in spontaneous speech despite not doing so in semantic fluency tasks.

Recently, Stojanovik and Ewijk (2008) pointed out that evidence from spontaneous speech is also problematic because conversational context is unconstrained. Some people with WS have been reported to hold specialised interests (Udwin, Howlin, & Davies, 1996), presumably with attendant specialist vocabulary that may emerge in constrained spontaneous speech (i.e., speech directed at topics of interest). Therefore, Stojanovik and Ewijk (2008) measured vocabulary diversity in a topic-constrained spontaneous speech task, with a WS group and control groups matched on receptive language and CA. The WS group produced neither more varied words nor more atypical words than the control groups, but, in fact, produced fewer of the lowest frequency words than controls.

There are hints from existing developmental studies indicating that lexical-semantic knowledge may be acquired atypically in WS. Stevens and Karmiloff-Smith (1997) argued that, when learning new words, children with WS were subject to the fast mapping and mutual exclusivity constraints, but not the taxonomic or whole-object constraints. Moreover,

Mervis and Bertrand (1997) found that children with WS learn words less referentially than typically-developing children, and that the stage of rapid vocabulary growth is less connected to semantic categorisation ability than it is in typical development. While Temple and colleagues (Temple et al., 2002) argue that such studies provide evidence that individuals with WS have an atypical lexical structure from the start of development, this need not be the case. For example, it is possible that atypical early learning processes have an impact on the type of lexical-semantic information encoded, with the lexicon structured normally. The former view attributes the use of unusual words in WS to a lexicon with atypical dynamics, the latter as a consequence of atypical learning processes.

Overall, our ability to assess the developing structure of the WS lexicon has been compromised by concerns about the limitations of the tasks used to assess it. Tyler and colleagues (Tyler et al., 1997) suggest that the lexical-semantic tasks that are difficult for participants with WS are often off-line tests that involve confounding task-demands. For example, while performance on semantic fluency tests might reflect the organisation of semantic information, it also requires retrieval strategies, which make metacognitive demands. In addition, although asking participants to define words yields information about semantic representations without requiring the participant to retrieve a name, it requires not only knowledge of the word's meaning, but also an understanding of what a "definition" is and how it is normally given; specifically, that it is a test of knowledge of the salient and diagnostic features of a category, to be listed in descending order of salience and diagnosticity. Poor performance on a definitions task, then, may arise from lexical-semantic problems but also from metacognitive demands (Benelli, Arcuri, & Marchesini, 1988). Thus, the WS group that performed similarly to a DS group on a definitions task in Bellugi and

colleagues' study (Bellugi et al., 1988) may have done so for metacognitive reasons, rather than for lexical-semantic ones.

The aim of the current study was to investigate whether there might be less information in the WS lexicon than suggested by vocabulary level. This is important for two reasons. First, it is important so that caregivers or teachers can interact with people with WS at a level appropriate to their intellectual ability, rather than the ability superficially suggested by their vocabulary. Second, it is important in order that theorists may accurately interpret what the presence of advanced vocabulary *per se* means for the developmental dissociation of cognitive abilities. Two paradigms were used to assess knowledge of the same semantic domain, with both participants with WS and typically developing controls. The first examined lexical-semantic processes with a definitions test; the second was a categorisation task in which participants were asked to sort objects into semantic categories. While the definitions task was expected to suffer from metacognitive confounds, the categorisation task was expected to minimise such confounding demands. However, it was necessary to conduct the definitions test in order to ascertain whether or not metacognitive concerns about the test were justified. If there were a convergence of performance on the two tests, it would suggest that the definitions task represents a valid measure of lexical-semantic knowledge.

Both paradigms focused on the same knowledge domain, animals, to facilitate cross-task comparison. Clearly, it is important to test with a domain in which individuals with WS are well versed, otherwise poor performance could be attributed to poor knowledge. Focusing on the domain of animals should maximise the chances of success at the tasks for the participants with WS, because it has been shown that individuals with WS as young as 10 have unimpaired basic knowledge in this area compared to verbal MA matched controls (Johnson & Carey, 1998).

Because WS is a developmental disorder, the data were analysed within a developmental framework. A developmental trajectories approach was adopted (Thomas, Annaz, Ansari, Scerif, Jarrold & Karmiloff-Smith, 2009). Functions of task performance and age are constructed, which allow developmental change to be compared across typically- and atypically-developing groups. Although longitudinal methods would ideally be used to investigate developmental change, a first approximation of developmental trajectories can derive from cross-sectional studies; these initial trajectories may subsequently be validated by longitudinal investigations. One benefit of employing trajectories that link performance on a task to a mental age measure is that they can be used to examine whether that performance is in line with the developmental state of other measures of cognitive ability, thereby assessing *developmental relations* within the atypical cognitive system. Although it is to be expected that people with WS will not perform at a CA-appropriate level, it may be that verbal mental-age normalises performance, which would show that typical developmental relations exist in the language systems of individuals with WS.

Method

Participants

There were two groups: 45 typically developing (TD) individuals and 15 individuals with a clinical diagnosis of Williams syndrome (WS) confirmed by the fluorescence in situ hybridisation (FISH) test. In a preliminary test session, the WS group was assessed on the British Picture Vocabulary Scale II (BPVS; Dunn, Dunn, Whetton, & Burley, 1997), a measure of receptive vocabulary. The WS group had a mean chronological age of 21;5 years (range = 12;0-44;11) and a mean vocabulary mental age of 9;7 years (range = 4;1-17;0). The vocabulary age for 3 older participants was at ceiling (17;0). Taking into account this ceiling (i.e., assigning all members of the WS group with CAs above 17;0 a CA of 17;0), the

standardised vocabulary test indicated a disparity between chronological and vocabulary age of 5 years and 11 months ($t(14) = 5.521, p < .001$). Receptive vocabulary was therefore clearly delayed in this group. The TD group had a mean chronological age of 7;6 years (range = 5;1-10;1) and a mean vocabulary mental age of 8;1 years (range = 5;1-13;1). Each participant consented to take part on the day of testing.

Materials

Definitions test

Using the Oxford Psycholinguistic Database (Wilson, 1988), 21 animals were selected for the participants to define, varying in frequency. A mixture of basic and superordinate category levels were selected. The animals chosen were *bird, fish, cat, lion, bee, elephant, ant, tortoise, spider, dolphin, kangaroo, crab, penguin, dinosaur, bat, beetle, whale, mammal, carnivore, reptile and marsupial*.

Categorisations Test

There were 20 categorisation questions in total. There were two rationales behind the choice of probe and animal grouping:

1. To investigate participants' responses to probes for properties not deducible from the available perceptual features
2. To examine the effect of perceptually similar distractors on these probes

Table 1 lists the categorisation questions.

===== *insert Table 1 about here* =====

Toy animals were used as stimuli (see Appendix 1 for the stimulus toys used for each categorisation question). These were kept covered during the definitions test so that participants could not base their definitions on the visual features of the toys.

Procedure

The definitions test was administered before the categorisation test.

Definitions test

Participants were asked, ‘What’s an x? Or can you tell me what an x is?’ If necessary, participants were also asked, ‘if you had to make another child understand what an x is, who did not know what it was, what would you say to help him?’ After the initial response, participants were prompted twice further for each animal with the phrase ‘Is there anything else you can tell me about x?’ The animals to be defined were presented in the same order for all participants. The superordinate definitions, such as ‘mammal’, ‘reptile’ or ‘marsupial’, were requested after the basic animal definitions, in order to avoid them priming participants’ responses.

The dependent measures were the number of correct features, and the number of salient and diagnostic features, given for each animal. Salient and diagnostic features were defined as those that allowed animals to be distinguished from each other or appear central to common definitions. This definition is necessarily subjective, because relevance of particular features for a particular goal (in this case, defining animals) is a matter of degree (Sperber & Wilson, 1987; Wilson & Sperber, 2004). To illustrate our use of these terms, an example of a feature that is both salient and diagnostic is *stripes* for a zebra. The fact that a zebra *has four legs* is salient but clearly not diagnostic. The fact that a frog *breathes through its skin while underwater* is diagnostic of frogs but not salient (or at least would not be expected to be

salient for our sample). In contrast, thematically or episodically related information would not be considered either salient or diagnostic (e.g. “I saw one on TV last night”, “It’s my brother’s favourite animal”).

Categorisations test

Participants were asked to group the animals according to the question asked. As a practice trial, a group of animals was placed on the table and the participants were asked to sort out which animals might be found in the circus. They were laid out in an order that did not correspond to the category question. As the animals were put out, the experimenter also named them. The category questions were administered in the same order for each participant. The dependent measure was the number of animals correctly placed within the probed category on each trial.

Results

Each of the following analyses is a repeated-measures ANCOVA with group as the between-subjects factor. The ANCOVA model included interaction terms between the verbal mental age (VMA) covariate and the between-subjects factor, to explore whether performance developed at a different rate in each group with respect to vocabulary ability (Thomas et al., 2009). The data were analysed with respect to VMA, rather than chronological age (CA), for two reasons. First, many studies have already established that lexico-semantic knowledge is not at CA-appropriate level in WS (e.g. Clahsen et al., 2004; Temple et al., 2002). Our participants with WS similarly demonstrated a delay in their receptive vocabulary ability compared to CA of around 6 years. Second, the key aim of the study is to investigate developmental relationships, namely whether lexico-semantic knowledge is commensurate

with vocabulary. It should be noted that three of the participants with WS were at ceiling (17;0 years) on the BPVS.

Definitions task

Correct features

There was a reliable positive relationship between VMA and the number of correct features given, $F(1,56) = 21.775, p < .001, \eta_p^2 = .280$, but no significant main effect of group, $F(1,56) = 0.449, p = .505, \eta_p^2 = .008$. There was, however, a reliable interaction of group and VMA, $F(1,56) = 4.753, p < .05, \eta_p^2 = .078$ (see Figure 1). Analysing the groups separately revealed that the number of correct features given reliably increased with VMA for the TD group, $F(1,43) = 23.497, p < .001, \eta_p^2 = .353$, but not the WS group, $F(1,13) = 2.559, p = .134, \eta_p^2 = .164$. At younger VMA, the TD and WS groups performed similarly, but there was evidence of divergence with faster development in the TD group thereafter. Overall, at higher vocabulary levels, definitions provided by individuals with WS were poorer than those of the TD controls.

===== insert Figure 1 about here =====

Salient and diagnostic features

The number of salient and diagnostic features reliably increased with VMA, $F(1,56) = 78.982, p < .001, \eta_p^2 = .585$, but there was no significant main effect of group, $F(1,56) = 1.050, p = .310, \eta_p^2 = .018$. However, performance improved with VMA faster in the TD group than in the WS group, $F(1,56) = 17.232, p < .001, \eta_p^2 = .235$ (see Figure 2). Again, at lower vocabulary abilities the performance of the groups overlapped but the trajectories then diverged with the performance of the TD group developing more quickly.

===== insert Figure 2 about here =====

Analysis of Errors

An error was defined as an incorrect feature offered for a given animal, and did not include omissions or irrelevant comments. The TD group's mean number of errors for each trial was 0.34 ($SD = 0.21$), the WS group's was 0.38 ($SD = 0.21$). Although the number of errors reliably decreased with increasing VMA, $F(1,56) = 6.456$, $p < .05$, $\eta_p^2 = .103$, there was no significant main effect of group, $F(1,56) < 0.001$, $p = .997$, $\eta_p^2 < .001$, nor interaction of group and VMA, $F(1,56) = 0.794$, $p = .377$, $\eta_p^2 = .014$. There was no fixed number of responses in this task, so the errors would not be expected to mirror the correct responses.

Feature analysis

The type of features produced was examined in more detail. Perceptual features were defined as those that could be known by recalling a mental image of a given animal (e.g., *has a beak*, *has big ears*); abstract features were those that could not be known from imagery of the animal alone (e.g., *is poisonous*, *lives underground*).

Overall, more perceptual features were given than abstract ones, $F(1,58) = 17.247$, $p < .001$, $\eta_p^2 = .229$. Although there was no reliable interaction of VMA and feature type for the TD group, $F(1,43) = 0.476$, $p = .494$, $\eta_p^2 = .011$, there was for the WS group, $F(1,13) = 6.935$, $p < .05$, $\eta_p^2 = .348$ (see Figure 3). While the WS group produced more abstract features with increasing VMA, $F(1,13) = 26.535$, $p < .001$, $\eta_p^2 = .671$, there was no significant change in the number of perceptual features, $F(1,13) = 0.200$, $p = .662$, $\eta_p^2 = .015$. The overall decrement in total features at higher receptive vocabulary levels therefore stems from the

absence of relevant perceptual detail rather than the properties of the animal (e.g., how it behaves, where it lives).

===== insert Figure 3 about here =====

Definitions task summary

Performance on the definitions task developed more slowly with VMA for the WS group than the TD group, measured by both the number of correct features and the number of salient and diagnostic features. However, this performance difference was not associated with any developmental group difference in errors. This implies that less semantic information is being acquired by the WS group, despite increasing vocabulary, or at least that less semantic information was elicited by this particular task.

Categorisation task

Correct categorisations

Performance on this measure improved reliably with VMA, $F(1,56) = 26.255, p < .001, \eta_p^2 = .319$, and the TD group scored significantly better than the WS group, $F(1,56) = 10.800, p < .01, \eta_p^2 = .162$. However, there was no reliable interaction of VMA and group, $F(1,56) = 0.311, p = .579, \eta_p^2 = .006$ (see Figure 4). The TD group performed better than the WS group at all VMA levels, but both groups developed at the same rate, with no divergence of the trajectories.

===== insert Figure 4 about here =====

Incorrect categorisations

Incorrect categorisations were defined as inclusion errors, i.e. animals incorrectly placed within the category probed by a given question. The TD group's mean number of errors for

each trial was 0.61 ($SD = 0.29$), the WS group's was 0.73 ($SD = 0.42$). The WS group made more of these errors than the TD group, $F(1,56) = 5.460$, $p < .05$, $\eta_p^2 = .089$, and there was a significant interaction of group and VMA, $F(1,56) = 16.419$, $p < .001$, $\eta_p^2 = .227$. While there was no reliable trend with VMA for such errors in the WS group, $F(1,13) = 0.507$, $p = .489$, $\eta_p^2 = .038$, the TD group showed a decrease in such errors with increasing VMA, $F(1,43) = 33.074$, $p < .001$, $\eta_p^2 = .435$. Therefore, the TD group tended to miscategorise less with increasing VMA, while the WS group retained the same level of errors across the VMA range.

Perceptual distractors

It was not possible to perform a meaningful feature analysis (as in the definitions task analysis), because no perceptual features were probed, as such. This is because perceptual information was readily available in the animal toys themselves. Instead, abstract features were probed, such as “Which live in a nest?” (See Appendix 1 for probes). However, it was still possible to investigate performance on trials that involved perceptual distractors (e.g. “Which are birds?”, where the response set was *eagle, swan, penguin, dragonfly* [with wings], *sea lion, horse, bat* [also with wings]). The TD group performed significantly better than the WS group on such trials, $F(1,56) = 4.399$, $p < .05$, $\eta_p^2 = .073$, and there was a reliable interaction of group and VMA, $F(1,56) = 15.017$, $p < .001$, $\eta_p^2 = .211$: while the TD group's performance improved reliably with increasing VMA, $F(1,43) = 24.137$, $p < .001$, $\eta_p^2 = .360$, the WS group's did not, $F(1,13) = 0.451$, $p = .513$, $\eta_p^2 = .034$ (see Figure 5).

===== insert Figure 5 about here =====

Categorisation task summary

For their level of receptive vocabulary, the TD group was better able to correctly categorise animals than the WS group. In addition, the WS group tended to make more errors than the TD group. The groups' ability to categorise correctly developed at a similar rate. However, as measured by performance on trials that involved perceptual distractors, abstract knowledge developed more slowly in the WS group than predicted by receptive vocabulary.

Cross-task comparison

In order to compare the performance of the WS group across tasks, it was necessary to make the measures used in each task comparable. One way to achieve this is to standardise the WS group's performance on the range of variability of the TD group (see Jarrold & Brock, 2004; Thomas, Annaz, Ansari, Scerif, Jarrold & Karmiloff-Smith, 2009). This standardisation generates Z-scores for each participant with WS, showing how far away performance is from that predicted by VMA, if the participant had been on the TD trajectory.

In order to directly compare performance on the definitions task with that on the categorisation task, participants' mean scores for each task were converted into Z-scores. Each participant's performance was standardised with respect to the distribution of scores for the typically developing children in each task. Figure 6 shows cross-task trajectories based on these Z-scores. Simple effects of task were independent of the covariate of VMA, because the covariate is a between-subjects factor, whereas task is a repeated-measures factor (see the following link for discussion of the use of repeated measures in ANCOVA: http://www.psyc.bbk.ac.uk/research/DNL/stats/Repeated_Measures_ANCOVA.html).

Therefore, these results are reported from an analysis that excludes the covariate (Degrees of freedom therefore differ between simple task effects and group effects or interactions). The TD group showed very similar relationships between each task and VMA, $F(1,43) = 0.054$, $p = .817$, $\eta_p^2 = .001$ (simple effects of task would be meaningless for the TD group, because the

mean Z-score for each task is zero by definition). The WS group's performance on the categorisation task was reliably worse than that their performance on the definitions task, $F(1,14) = 13.109$, $p < .01$, $\eta_p^2 = .484$, though there was no significant difference in the gradients of the trajectories, $F(1,13) = 1.386$, $p = .260$, $\eta_p^2 = .096$. Figure 6 also captures the divergence of the WS trajectory from the TD trajectory with increasing VMA, while the category trajectory runs parallel.

===== insert Figure 6 about here =====

As a final comparison between the definitions and categorisation tasks, performance with 'elephant' was examined in each, using the features probed in the categorisation task: 'where it lives', 'what it eats', 'source of ivory', 'is a mammal', and 'lives for a long time'.

'Elephant' was chosen for this analysis because it had the highest number of features probed in the categorisation task that also appeared in the definitions task. Figure 7 demonstrates that the features related to elephants that were probed in the categorisation task were more often successfully responded to in this task than they were produced in the definitions task, $F(1,58) = 200.192$, $p < .001$, $\eta_p^2 = .775$. While there was no reliable group difference in how often these features were produced in the definitions task, $F(1,58) = 1.697$, $p = .198$, $\eta_p^2 = .028$, the TD group successfully categorised the elephant on the basis of these features significantly more often than the WS group did, $F(1,58) = 18.253$, $p < .001$, $\eta_p^2 = .239$ (see Figures 7a and 7b).

===== insert Figures 7a and 7b about here =====

Discussion

The aim of the study was to investigate the relationship between lexico-semantic knowledge and vocabulary in WS compared to a TD control group. Two tasks were used to measure lexico-semantic knowledge: a definitions task, which was expected to make additional metacognitive demands, and a categorisation task, which was considered a purer test of lexico-semantic knowledge. The results indicated that the WS group's performance on the definitions task began at a level commensurate with VMA, but diverged from that of the typically developing group. The WS group's performance on the categorisation task developed at a similar rate to that of the typically developing participants, but was markedly poorer on average than predicted by VMA. This surprising pattern of results indicates that individuals with WS have *less* lexico-semantic knowledge than expected given their level of receptive vocabulary, although this knowledge appears to develop in step with vocabulary at this lower level. These results also suggest that metacognitive concerns about the definitions task may be well-founded, a point to which we return below.

The WS group's disparity of lexico-semantic knowledge and receptive vocabulary may be due, at least in part, to poorly-delineated semantic categories: the WS group made more categorisation errors than the TD group, with the effects of VMA accounted for. Moreover, while the TD group made fewer such errors with increasing VMA, presumably reflecting increasingly well-defined category boundaries, no such trend was evident for the WS group. These results are in line with a previous finding that individuals with WS acquire additional lexico-semantic information throughout development, without accompanying conceptual change. Johnson and Carey (1998) suggested that cocktail party syndrome in WS results from adequate 'enrichment' learning processes in the absence of analytic and metacognitive skills. Udwin and Yule (1990) had found that about a third of a sample of 43 children with WS met the criteria for cocktail party syndrome, which include well-formed speech that nevertheless

lacks communicative content, along with frequent use of conversational fillers and stereotypical social phrases. Johnson and Carey predicted a dissociation between general knowledge of animals (e.g., number of legs, what it eats, where it lives) and core folk-biological concepts (e.g., the determinants of species identity, the notion that humans are one animal of many). They tested intuitive biological knowledge in WS and two TD groups, one matched on VMA and the other a non-matched group of 6-year-olds. The WS group performed similarly to the VMA-matched group on a test battery for biological general knowledge, but performed significantly worse on a test battery for folk-biological concepts thought to be acquired between the ages of six and twelve, and at a similar level to the 6-year-olds. The authors concluded that the WS group had not acquired folk-biological concepts appropriate for VMA, even though the requisite general knowledge was probably in place¹.

A recent study of comprehension of non-literal similarity provides a parallel with Johnson and Carey's results. Thomas et al. (submitted) administered a simple picture-based categorisation task to individuals with WS and children between 5 and 11. The results indicated that the individuals with WS understood both perceptual similarity across category boundaries (e.g., an orange is perceptually similar to the Sun) and also functional similarity across category boundaries (e.g., an oven and the Sun both heat things up). However, in contrast to the TD group, the WS group did not develop a preference for functional similarity over perceptual similarity in comparison judgements, consistent with the notion that individuals with WS do not develop a conceptual structure that flexibly utilises functional relations, despite showing clear evidence that functional relations themselves have been encoded. This is consistent with Johnson and Carey's finding that the WS group demonstrated evidence of the requisite pieces of knowledge on which to base functional

categories but not evidence of functionally-organised *categories* which would be necessary to make similarity judgements. Taken together with the results of the current study, it seems reasonable to suggest that individuals with WS have poorer *organisation* of knowledge than predicted by the *amount* of knowledge they have accrued.

One puzzling aspect of our results is that in the definitions task, while both groups gave more abstract features with increasing VMA, and the TD group also produced more perceptual features, the WS group demonstrated no such increase in production of perceptual features. This appears to be because the youngest participants were particularly unlikely to produce abstract features, rather than because the older children were particularly likely to do so. In the categorisation task, the WS group performed more poorly than the TD group on categorisations that involved perceptual distractors (e.g. “Which are birds?” with dragonfly and bat among the distractors). Furthermore, while the TD group’s performance on such trials improved with VMA, the WS group showed no mental-age-related trend. This suggests that the participants with WS found perceptual features particularly salient, which may go some way to explaining why abstract features were offered less often by younger participants with WS in the definitions task.

In order to further compare performance across tasks, additional analyses were conducted, focusing on one of the animals probed in the study, elephant, on a feature-by-feature basis. The features of ‘elephant’ probed in the categorisation tasks were ‘where it lives’, ‘what it eats’, ‘source of ivory’, ‘is a mammal’, and ‘lives for a long time’; these same features were produced by participants in the definitions task. These salient and diagnostic features were very rarely offered in the definitions task, despite the fact that such knowledge was often demonstrated in the categorisation task. This pattern of results reflects the relative insensitivity of the definitions task for assessing knowledge of salient and diagnostic features.

In order to perform well on a definitions task, the participant must list features in descending order of salience and diagnosticity. However, the participant must also understand that this is what is required for task success. It is not clear that younger typically developing children or individuals with developmental disorders will understand this, or would even be helped by careful explanation, given the metacognitive nature of understanding and monitoring task success. While there was no group difference in how often these features were produced in the definitions task, the TD group successfully *categorised* the elephant on the basis of these features more often than the WS group did. This pattern of results repeats the message from the main analyses: the participants with WS demonstrated lexico-semantic knowledge that was poorer than predicted by receptive vocabulary, but developed at a similar rate to that observed in typical development.

Another, more tentative, way of comparing performance across tasks is to contrast the salient and diagnostic features given in the definitions task with correct categorisations, given that categorisations were always made on the basis of salient and diagnostic features. Although, at lower vocabulary abilities, the performance of the groups was similar, the performance of the TD group developed more quickly than that of the WS group. In contrast, the WS group were stably poorer than the TD group across the entire developmental trajectory in the categorisation task. Whether comparing the categorisation task to the definitions task in terms of total correct features or salient and diagnostic ones, different patterns of performance were seen across the two tasks, indicating that the tasks were measuring different abilities, consistent with the suggestion that asking participants to produce definitions assesses metacognitive skills in addition to lexico-semantic knowledge (cf. Benelli et al., 1988).

Finally, the findings of the current study are in line with those of Temple and colleagues (Temple et al., 2002; Clahsen, Ring, & Temple, 2004), who found that individuals with WS

performed worse at a version of the BPVS with more semantic distractors than typically developing controls matched on MA. Temple and colleagues suggested that the standard version of the BPVS may overestimate lexico-semantic knowledge in WS because, through its forced choice design (i.e., which one of four pictures goes with the named word), decisions can be made without possessing full knowledge of the word meaning. The results of the current study support this conclusion. In the absence of some ‘gold standard’ measure of language ability, it is not possible to definitively state whether lexico-semantic knowledge is worse than other areas of language in WS. It is possible that lexico-semantics only appears to be out of step with vocabulary in WS because the BPVS is a poor measure of vocabulary for some populations of individuals with learning difficulties. As with many standardised tests, the BPVS is far from ‘pure’, but involves many non-central task demands, such as holding the task aim in mind, inhibiting responding on the basis of simply liking a picture, and the ability to generalise from tokens to types. The possible limitations of the BPVS for atypical populations point to the same conclusion as the current data. Assertions of exceptional lexical knowledge in Williams syndrome should be treated with caution.

Acknowledgements

This research was supported by European Commission grant NEST-029088(ANALOGY), British Academy grant SG30602, and MRC grant G0300188.

References

- Anderson, M. (1998). Mental retardation general intelligence and modularity. *Learning and individual differences, 10*, 159-178.
- Ansari, D., Donlan, C., Thomas, M. S. C., Ewing, S. A., Peen, T., & Karmiloff-Smith, A. (2003). What makes counting count? Verbal and visuo-spatial contributions to typical and atypical number development. *Journal of Experimental Child Psychology, 85*, 50-62.
- Bellugi, U., Bihrlle, A., Jernigan, T., Trauner, D., & Doherty, S. (1990). Neuropsychological, neurological, and neuroanatomical profile of Williams syndrome. *American Journal of Medical Genetics Supplement, 6*, 115–125.
- Bellugi, U., Bihrlle, A., Neville, H., Jernigan, T., & Doherty, S. (1992). Language, cognition, and brain organization in a neurodevelopmental disorder. In M. Gunnar & C. Nelson (Eds.), *Developmental Behavioral Neuroscience* (pp. 201-232). Hillsdale, NJ: Lawrence Erlbaum Associates.
- Bellugi, U., Lichtenberger, L., Jones, W., Lai, Z., & St. George, M. (2000). The neurocognitive profile of Williams syndrome: A complex pattern of strengths and weaknesses. *Journal of Cognitive Neuroscience, 12:S*, 7-29.
- Bellugi, U., Marks, S., Bihrlle, A., & Sabo, H. (1988). Dissociation between language and cognitive function in Williams Syndrome. In D. Bishop & K. Mogford (Eds.), *Language development in exceptional circumstances*. Edinburgh, U.K.: Churchill Livingstone.
- Bellugi, U., Wang, P. P., & Jernigan, T. L. (1994). Williams syndrome: An unusual neuropsychological profile. In S. Broman & J. Grafman (Eds.), *Atypical cognitive deficits in developmental disorders: Implications for brain function*. Hillsdale, NJ: Erlbaum.

- Benelli, B., Arcuri, L., & Marchesini, G. (1988) Cognitive and linguistic factors in the development of word definitions. *Journal of Child Language*, 15, 619-635.
- Brock, J. (2007). Language abilities in Williams syndrome: a critical review. *Development and Psychopathology*, 19, 97-127.
- Brock, J., Jarrold, C., Farran, E. K., Laws, G., & Riby, D. (2007). Do children with Williams syndrome have really good vocabulary knowledge? Methods for comparing cognitive and linguistic abilities in developmental disorders. *Clinical Linguistics & Phonetics*, 21, 673-688.
- Clahsen, H., Ring, M. & Temple, C. (2004). Lexical and morphological skills in English-speaking children with Williams Syndrome. In S. Bartke & J. Siegmüller (Eds.), *Williams Syndrome across Languages* (pp.221-244). Benjamins: Amsterdam.
- Donnai, D., & Karmiloff-Smith, A. (2000). Williams syndrome: From genotype through to the cognitive phenotype. *American Journal of Medical Genetics*, 97, 164-171.
- Dunn, L. M., Dunn, L. M., Whetton, C., & Burley, J. (1997). *British Picture Vocabulary Scale II*. Windsor, U.K.: NFER-Nelson.
- Ewart, A. K., Morris, C. A., Atkinson, D., Jin, W., Sternes, K., Spallone, P., Stock, A. D., Leppert, M., & Keating, M. T. (1993). Hemizyosity at the elastin locus in an developmental disorder, Williams Syndrome. *Nature Genetics*, 5, 11-16.
- Farran, E. K., & Jarrold, C. (2003). Visuo-spatial cognition in Williams syndrome: Reviewing and accounting for strengths and weaknesses in performance. *Developmental Neuropsychology*, 23, 173-200.

- Howlin, P., Davies, M., & Udwin, O. (1998). Cognitive functioning in adults with Williams syndrome. *Journal of Child Psychology and Psychiatry*, *39*, 183-189.
- Jarrold, C., & Brock, J. (2004). To match or not to match? Methodological issues in autism-related research. *Journal of Autism and Developmental Disorders*, *34*, 81-86.
- Jarrold, C., Hartley, S. J., Phillips, C., & Baddeley, A. D. (2000) Word fluency in Williams syndrome: Evidence for unusual semantic organization. *Cognitive Neuropsychiatry*, *5*, 293-318.
- Johnson, S. C. & Carey, S. (1998). Knowledge enrichment and conceptual change in folkbiology: evidence from Williams syndrome. *Cognitive Psychology*, *37*, 156-200.
- Karmiloff-Smith, A. (1998) Development itself is the key to understanding developmental disorders. *Trends in Cognitive Sciences*, *2*, 389-398.
- Laing, E., Grant, J., Thomas, M., Parmigiani, C., Ewing, S., & Karmiloff-Smith, A. (2005). Love is...an abstract word: The influence of lexical semantics on verbal short-term memory in Williams syndrome. *Cortex*, *41*, 169-179.
- Mervis, C., & Bertrand, J. (1997). Developmental relations between cognition and language: Evidence from Williams syndrome. In L. B. Adamson & M. A. Romski (Eds.), *Research on communication and language disorders: Contribution to theories of language development* (pp.75-106). New York: Brookes.
- Morris, C. A., Dempsey, S. A., Leonard, C. O., Dilts, C., & Blackburn, B. L. (1988). Natural history of Williams syndrome: Physical characteristics. *Journal of Paediatric Medicine*, *113*, 318-326

- Rossen, M., Klima, E. S., Bellugi, U., Bihrlé, A., & Jones, W. (1996). Interaction between language and cognition: Evidence from Williams syndrome. In J. H. Beitchman, N. J. Cohen, M. M. Konstantareas & R. Tannock, *Language learning and behavior* (pp. 367-92). New York: Cambridge University Press.
- Sperber, D. & Wilson, D. (1987) *Precis of Relevance: Communication and Cognition. Behavioral and Brain Sciences. 10, 697-754*
- Stevens, T. & Karmiloff-Smith, A. (1997) Word learning in a special population: Do individuals with Williams syndrome obey lexical constraints? *Journal of Child Language, 24, 737-765.*
- Stojanovik, V., & van Ewijk, L. (2008). Do children with Williams syndrome have unusual vocabularies? *Journal of Neurolinguistics, 21, 18-34.*
- Temple, C. M., Almazan, M. & Sherwood, S. (2002) Lexical skills in Williams' syndrome: A cognitive neuropsychological analysis. *Journal of Neurolinguistics, 15, 463-495.*
- Thal, D., Bates, E., & Bellugi, U. (1989). Language and cognition in two children with Williams Syndrome. *Journal of Speech and Hearing Research, 3, 489-500.*
- Thomas, M. S. C. (2008). L'acquisition du langage dans les pathologies du développement [Language development in developmental disorders]. In M. Kail, M. Fayol, & M. Hickmann, *L'apprentissage des langues*, (pp. 451-475). Paris: CNRS Editions.
- Thomas, M. S. C., Annaz, D., Ansari, D., Scerif, G., Jarrold, C., & Karmiloff-Smith, A. (2009). Using developmental trajectories to understand developmental disorders. *Journal of Speech, Language, and Hearing Research, 52, 336-358.*

- Thomas, M. S. C., Dockrell, J. E., Messer, D., Parmigiani, C., Ansari, D., & Karmiloff-Smith, A. (2006). Speeded naming, frequency and the development of the lexicon in Williams syndrome. *Language and Cognitive Processes, 21*, 721-759.
- Thomas, M. S. C., & Karmiloff-Smith, A. (2005). Can developmental disorders reveal the component parts of the human language faculty? *Language Learning and Development, 1*, 65-92.
- Thomas, M. S. C., Purser, H. R. M., & Richardson, F. M. (in press). Modularity and developmental disorders. In: P. D. Zelazo (Ed), *Oxford Handbook of Developmental Psychology*. Oxford: Oxford University Press.
- Thomas, M. S. C., van Duuren, M., Purser, H. R. M., Mareschal, D., Ansari, D., & Karmiloff-Smith, A. (submitted). The development of metaphorical language comprehension in typical development and in Williams syndrome.
- Tyler, L. K., Karmiloff-Smith, A., Voice, K., Stevens, T., Grant, J., Udwin, O., Davies, M., Howlin, P. (1997) Do individuals with Williams syndrome have bizarre semantics? Evidence for lexical organization using an on-line task. *Cortex, 33*, 515-527.
- Udwin, O. & Dennis, J. (1995). Psychological and behavioural phenotypes in genetically determined syndromes: A review of research findings. In G. O'Brien and W. Yule (Eds.) *Behavioural Phenotypes. Clinics in Developmental Medicine* No. 138. London: MacKeith Press.
- Udwin, O., & Yule, W. (1990). Expressive language of children with Williams syndrome. *American Journal of Medical Genetics Supplement, 6*, 108-114.

Volterra, V., Caselli, M.C., Capirci, O., Tonucci, F., Vicari, S. (2003). Early linguistic abilities in Italian children with Williams Syndrome. *Developmental Neuropsychology*, 23, 33-58.

Wilson, D. & Sperber, D. (2004). Relevance Theory. In L. Horn & G. Ward (Eds.) *The Handbook of Pragmatics* (pp. 607-632). Oxford: Blackwell.

Wilson, M. D. (1988). The MRC Psycholinguistic Database: Machine Readable Dictionary, Version 2. *Behavioural Research Methods, Instruments and Computers*, 20, 6-11.

Footnotes

¹ While no task in the biological general knowledge battery appeared to make particular metacognitive demands, two of the five tasks of the folk-biological concepts battery may have done so (*Death*, as part of which participants were asked several open-ended questions about death, such as “What happens to a person when they die?”, and *Species Transformations*, in which stories were told of animals being transformed to look like other animals [e.g. a tiger into a lion], either by dressing-up or by surgery, and participants were asked, e.g., “Is it a tiger or a lion?”). Despite these concerns, the same basic pattern of results held across all five tasks of the folk-biological concepts battery.

Table 1.

Categorisation questions and whether perceptual distractors were used in each case

Categorisation question	Perceptual distractors?
Which would you find in the sea?	N
Which are birds?	Y
Which are insects?	Y
Which are farm animals?	Y
Which are jungle animals?	N
Which can fly?	N
Which eat meat/fish vs. grass/vegetation?	N
Which live in hot places vs. cold places?	N
Which can swim well?	N
Which live in a nest?	N
Which make ivory?	N
Which can sting?	Y
Which can lay eggs?	N
Which are the two biggest in real life vs. two smallest?	Y

Which are rare and which are common animals? N

Which are reptiles? N

Which are mammals? N

Which ones live to be very old? N

Is a penguin the same kind of thing as a sea lion or eagle? Y

Is an octopus the same kind of thing as a jellyfish or spider? Y

Figures

Figure 1. Mean number of correct features given by participants in the definitions task plotted against verbal mental age in years. WS = Williams syndrome, TD = typically developing.

Figure 2. Mean number of salient/diagnostic features given by participants in the definitions task plotted against verbal mental age in years. WS = Williams syndrome, TD = typically developing.

Figure 3. Mean number of perceptual and abstract features given by participants in the definitions task plotted against verbal mental age in years. WS = Williams syndrome, TD = typically developing. Note the low R^2 value for the TD perceptual trajectory reflects wide variation despite an overall increase with age.

Figure 4. Mean number of correct categorisations plotted against verbal mental age in years. WS = Williams syndrome, TD = typically developing.

Figure 5. Mean number of correct categorisations on trials with perceptual distractors plotted against verbal mental age in years. WS = Williams syndrome, TD = typically developing.

Figure 6. Z-scores by group and task plotted against verbal mental age in years. WS = Williams syndrome, TD = typically developing.

Figure 7a. Mean number of correct features given by participants for 'elephant' in the definitions task plotted against verbal mental age in years. WS = Williams syndrome, TD = typically developing.

Figure 7b. Mean number of correct categorisations for 'elephant' plotted against verbal mental age in years. WS = Williams syndrome, TD = typically developing.

Figure 1

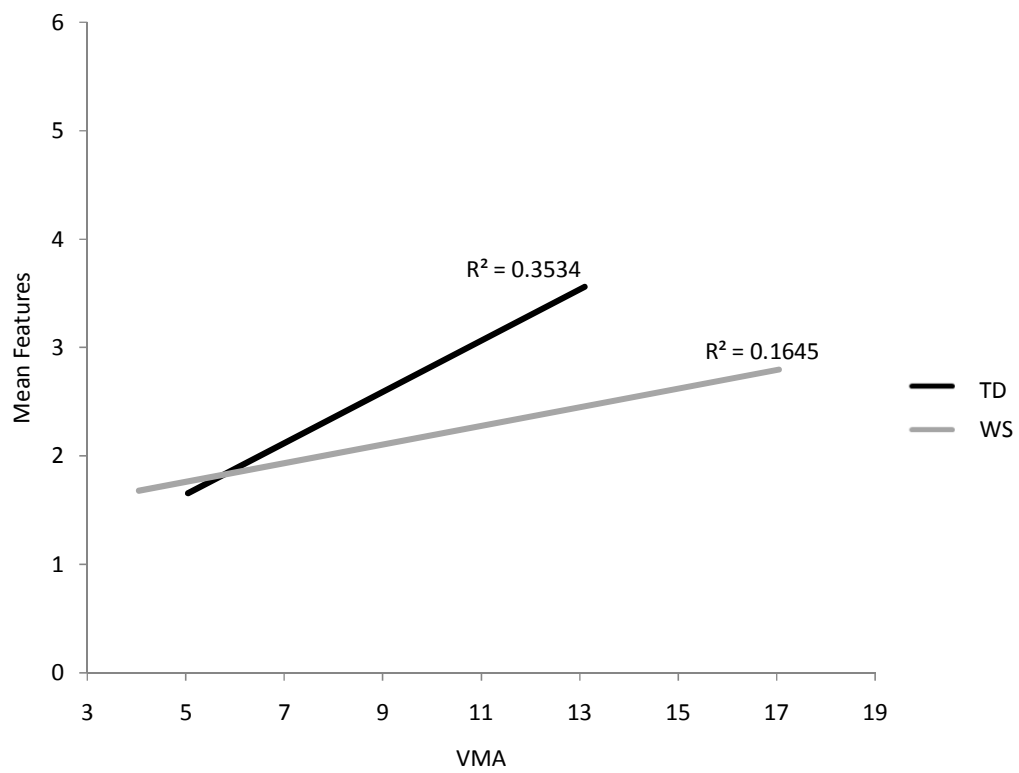


Figure 2

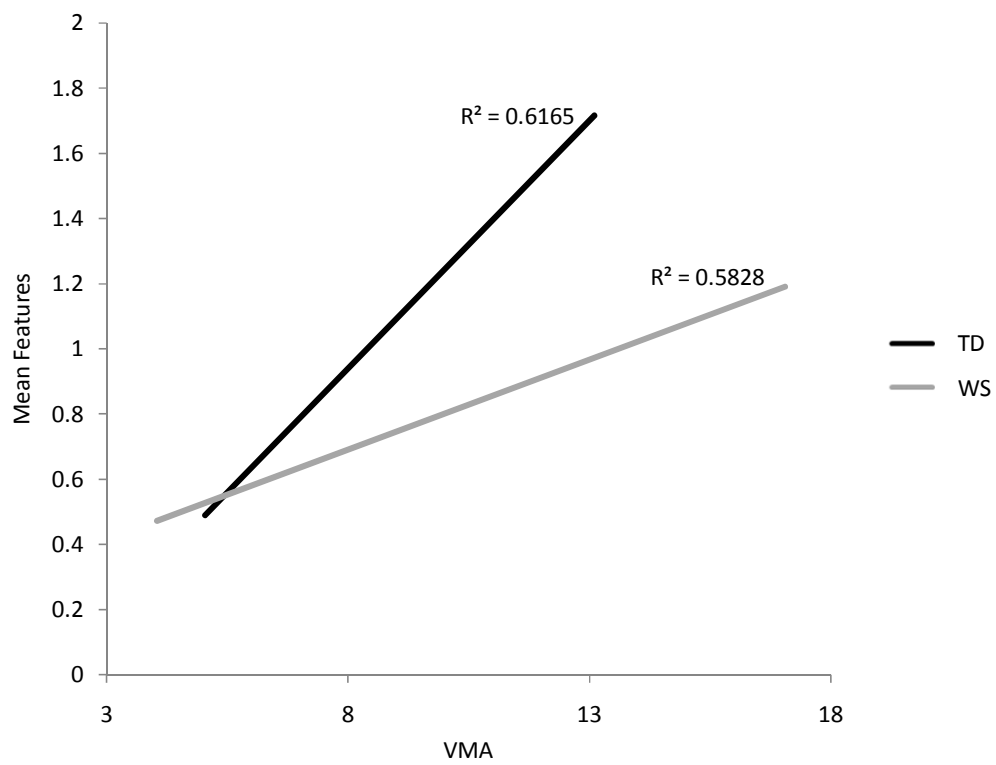


Figure 3

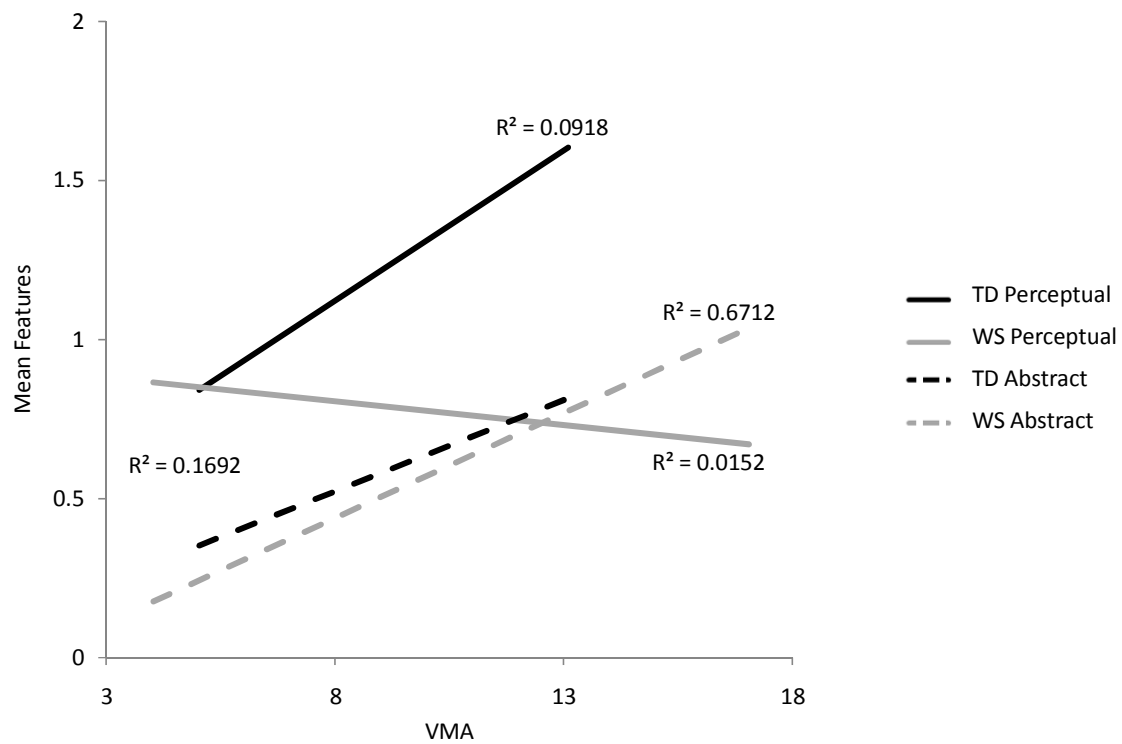


Figure 4

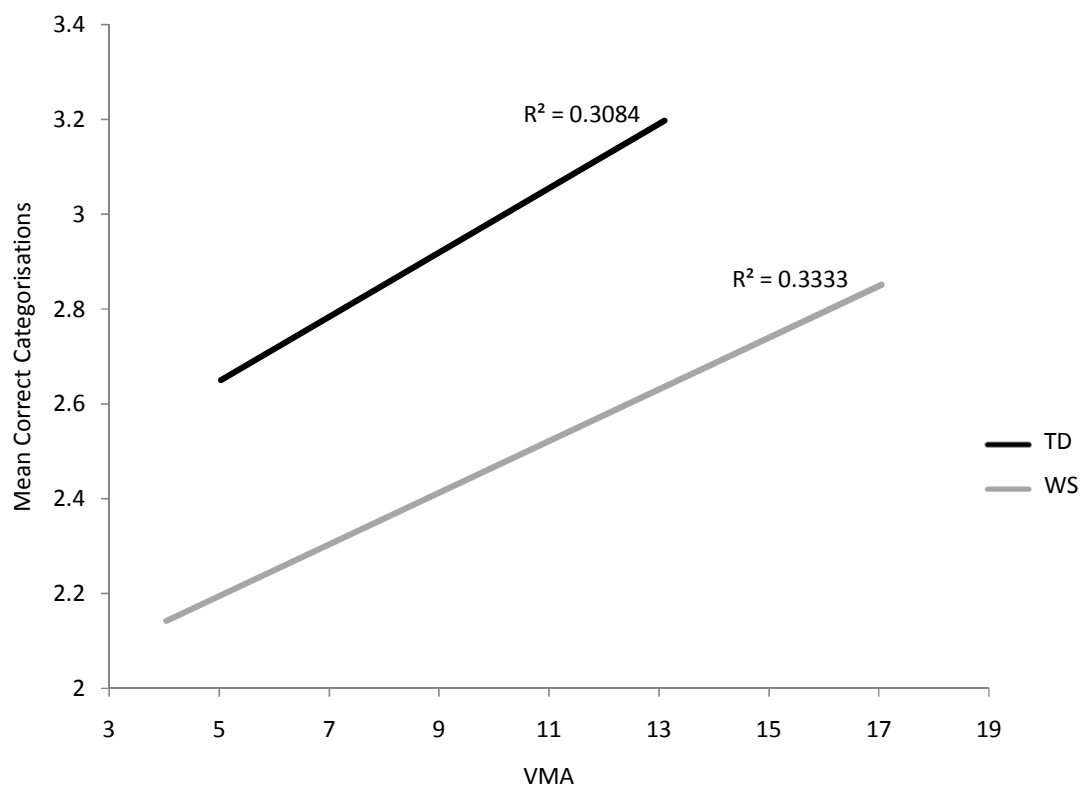


Figure 5

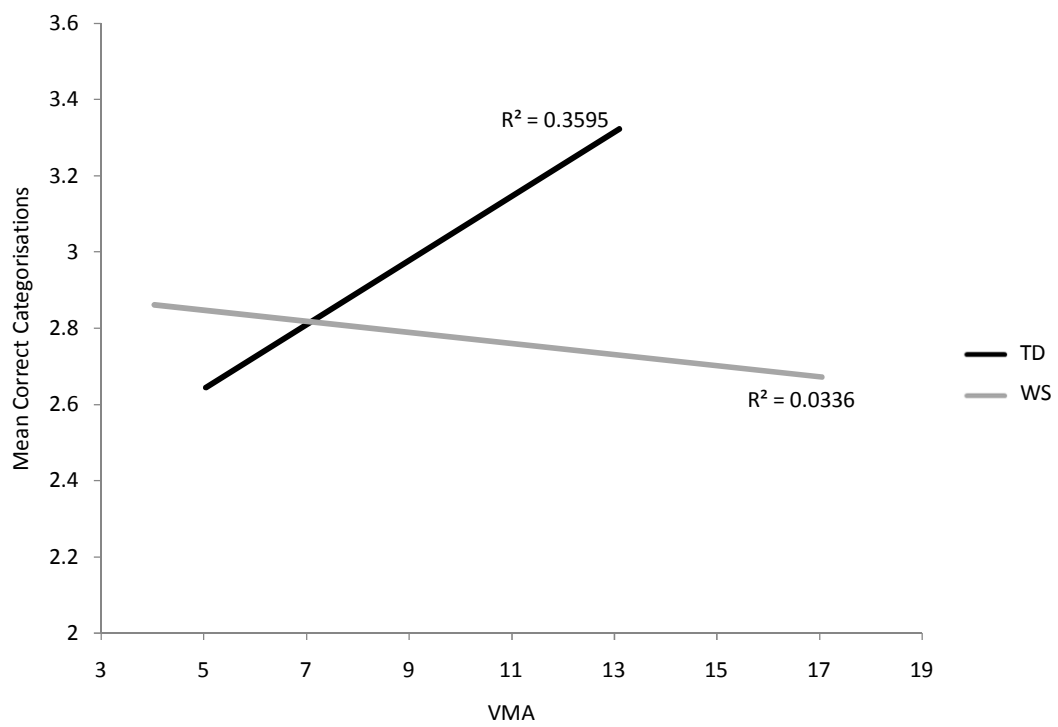


Figure 6

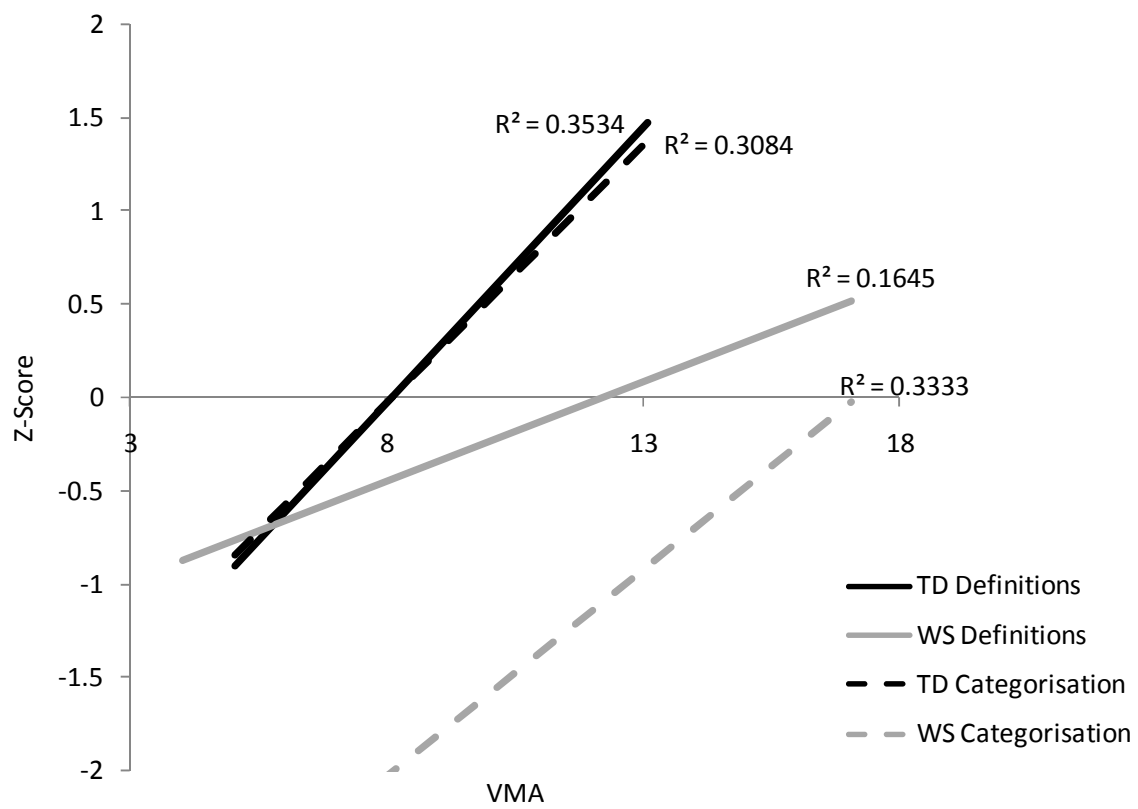


Figure 7a

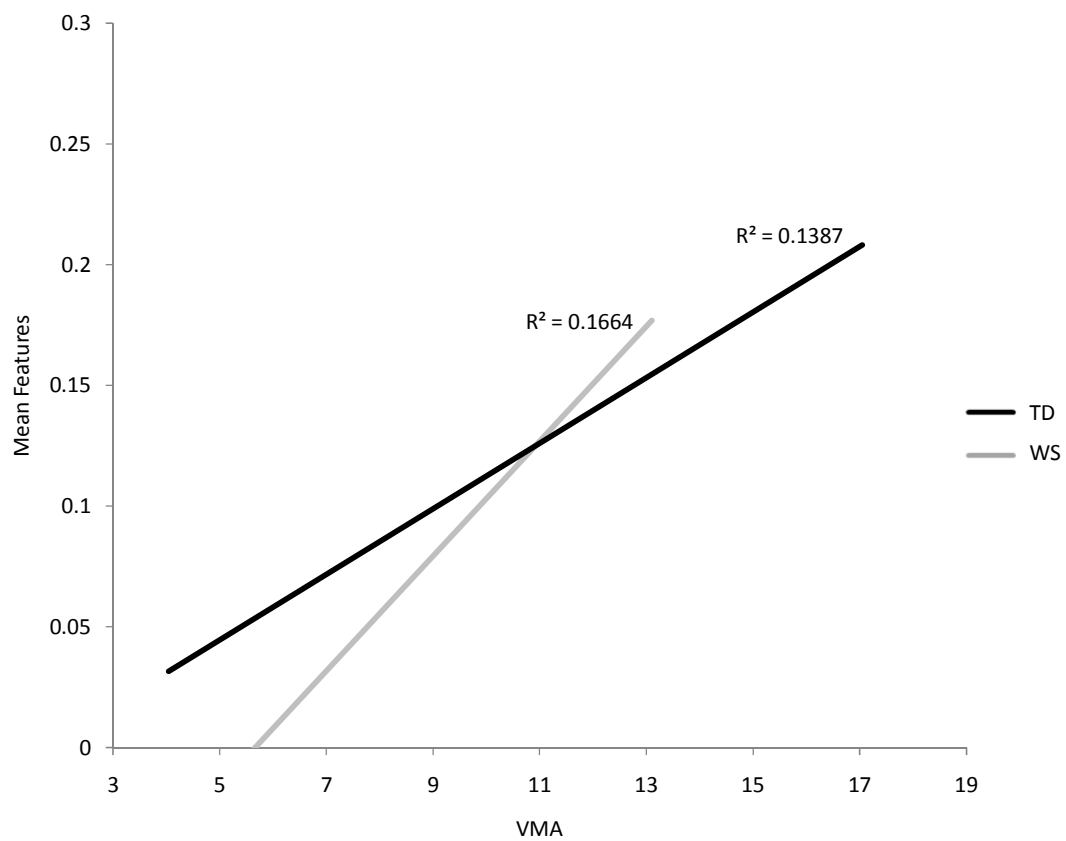
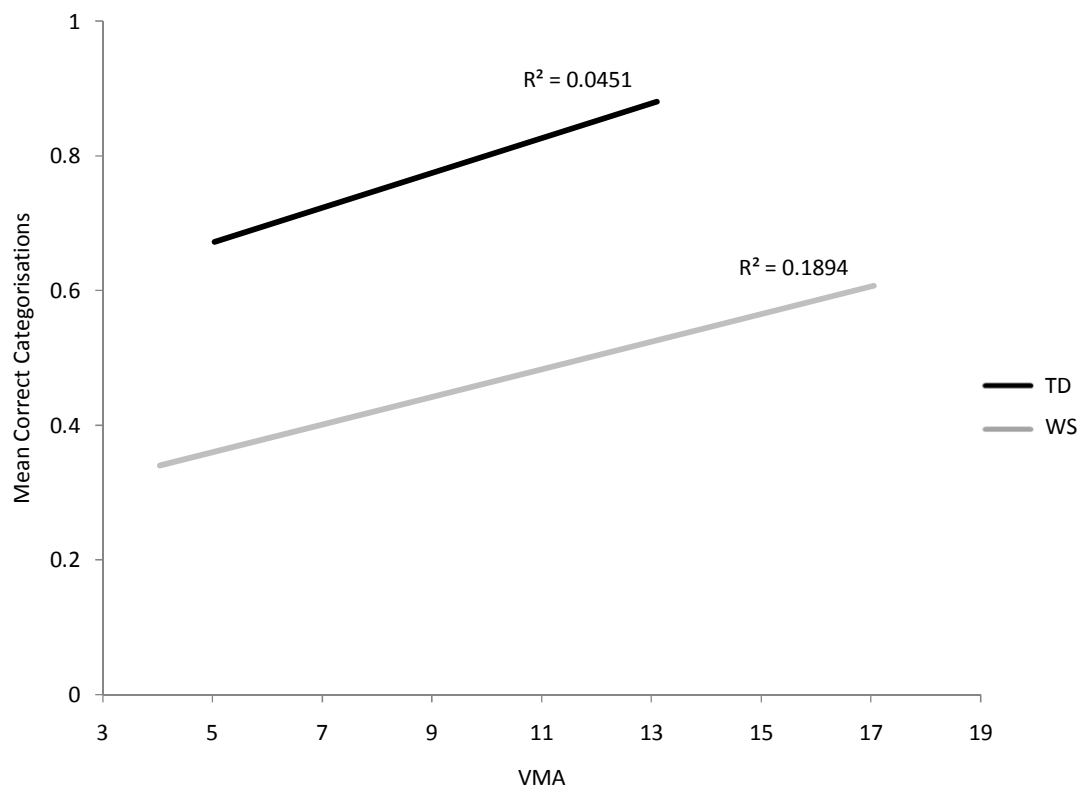


Figure 7b



Appendix 1. Stimulus animal toys used in the categorisation task, by question

Categorisation question	Within category	Outside category
Which would you find in the sea?	Crab, octopus, sealion, whale, jellyfish	Frog, platypus, crocodile, stag, lion
Which are birds?	Eagle, swan, penguin	Dragonfly, sealion, horse, bat
Which are insects?	Bee, dragonfly, beetle, ant	Octopus, crab, calf, frog
Which are farm animals?	Pig, cow, horse, goat, ram	Zebra, cheetah, stag, swan
Which are jungle animals?	Elephant, lion,	Sealion, horse, bear,

	panther,	penguin
	rhino	
Which can fly?	Bee, eagle,	Jellyfish,
	beetle, bat	spider,
		penguin, pig
Which eat meat/fish vs. grass/vegetation?	Whale,	Elephant,
	panther,	pig, goat
	bear	
Which live in hot places vs. cold places?	Zebra,	Bear,
	snake	penguin
Which can swim well?	Penguin,	Rhino, pig,
	dolphin,	cheetah,
	octopus,	bear, horse
	crocodile,	
	frog	
Which live in a nest?	Swan, bee,	Crab, cat,
	ant	calf
Which make ivory?	Elephant,	Crocodile,
	rhino	dolphin
Which can sting?	Bee,	Octopus,
	jellyfish	beetle

Which can lay eggs?	Swan, eagle, crocodile, T-rex, Frog	Elephant, dolphin, pig, bat, ram
Which are the two biggest in real life vs. two smallest?	Elephant, whale, pig, dog	Tortoise, spider, crab, bat, ant
Which are rare and which are common animals?	Panther, whale, eagle	Pig, ant, horse
Which are reptiles?	T-rex, snake, crocodile, tortoise	Sealion, platypus, eagle, jellyfish
Which are mammals?	Elephant, bat, whale, lion	Frog, octopus, eagle, beetle
Which ones live to be very old?	Tortoise, elephant, chimp	Frog, swan, ant
Is a penguin the same kind of thing as a sea lion or eagle?	Eagle	Sealion

Is an octopus the same kind of thing as a jellyfish or Jellyfish Spider
spider?
